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Ultrastructure of the Wall of the Dandy-Walker Cyst

A Case Report

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Summary. The ultrastructure of the wall of the Dandy-Walker cyst has been described rarely. A boy aged 2 years was confirmed clinically, neuroradiologically, and operatively as having a Dandy-Walker cyst in the posterior fossa. The cyst wall obtained during surgery consisted of an outer arachnoid cell layer, intermediate interwoven neuroglial strands, and an inner layer of cells which lacked the characteristic appearance of ependyma. An unusual finding was a small, buried island of ependymal cells in the intermediate layer of the neuroglial tissue. Ultrastructural study of the cyst wall provides a better understanding of the pathogenesis of the Dandy-Walker syndrome.

Key words: Dandy-Walker syndrome – Dandy-Walker cyst – Ependymal cell – Ultrastructure – Electron microscopy

Introduction

Hydrocephalus as a result of congenital atresia of the foramina of Luschka and Magendie of the fourth ventricle was described first by Dandy and Blackfan [3] and then evaluated by Taggart and Walker [13]. The "Dandy-Walker syndrome" was designated by Benda [1] as a specific type of obstructive hydrocephalus, the so-called "atresia of the foramen Magendie". The name Dandy-Walker syndrome was apparently given to parallel the Arnold-Chiari syndrome. Dandy-Walker syndrome consists of a group of hindbrain malformations characterized by hypoplasia of the cerebellar vermis, enlargement of the fourth ventricle into a posterior fossa cyst, and, usually, hydrocephalus. Raimondi et al. [11] considered the basic clinical problem to be the presence of a posterior fossa cyst which is entirely within the confines of the fourth ventricle, and which causes complete occlusion of the foramina of Luschka and Magendie. They referred to this clinical entity as the Dandy-Walker cyst.

There have been various descriptions based on light-microscopic observations of the cyst wall in this syndrome [2, 4, 6–8, 10] but no electron-microscopic observations have been reported. In this paper, we demonstrate the ultrastructure of the wall of a posterior fossa cyst, which was neuroradiologically and operatively diagnosed as a Dandy-Walker cyst.

Case Report

A boy of 2 years was admitted with progressive hydrocephalus to our neurosurgical clinic in August 1982. X-rays of the skull showed

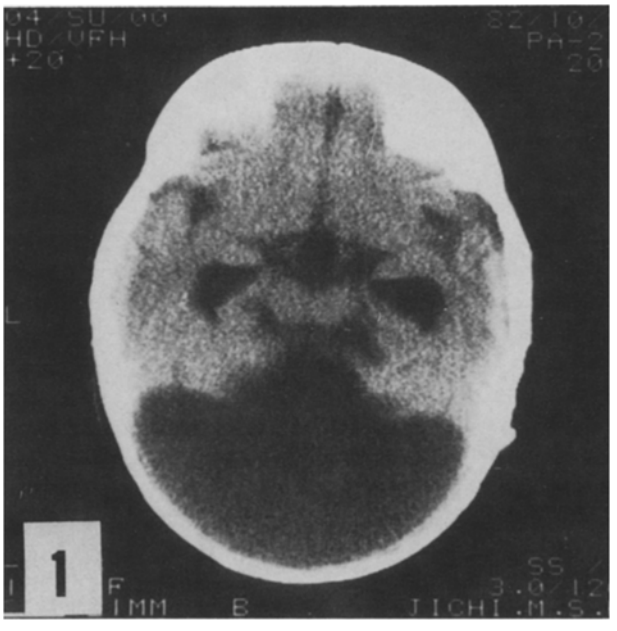

Fig. 1. A plain CT scan shows a large low density cyst confined within the fourth ventricle and associated with hypolasia of the cerebellar vermis, and is compatible with a Dandy-Walker cyst

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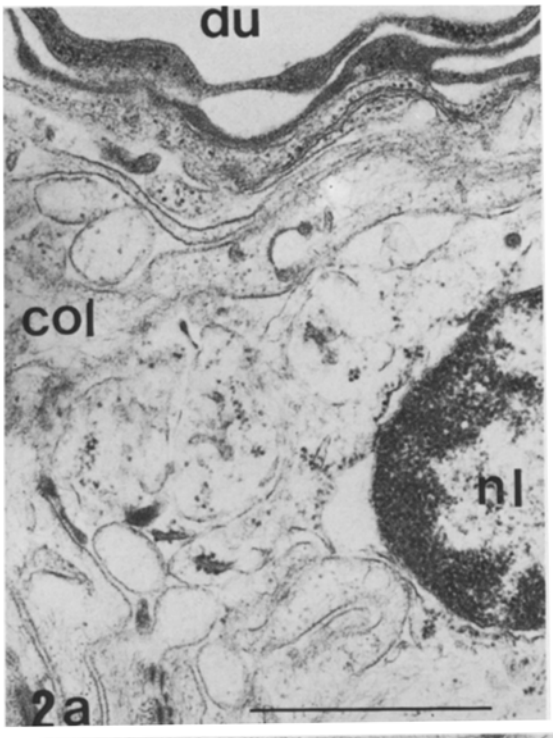
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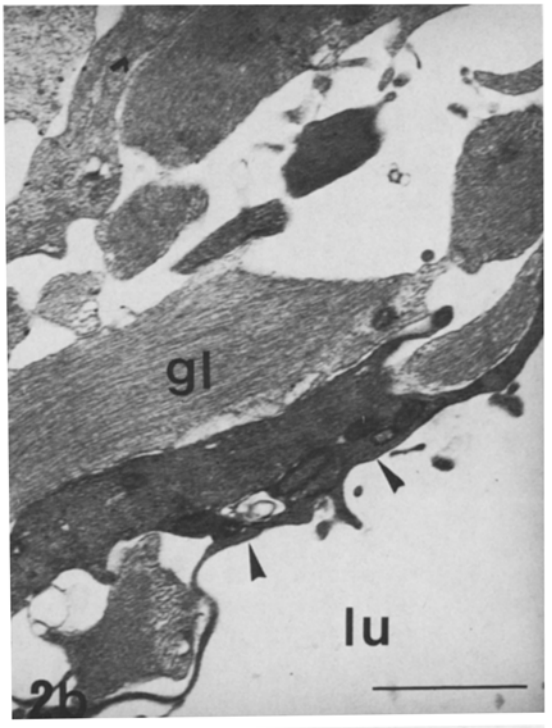
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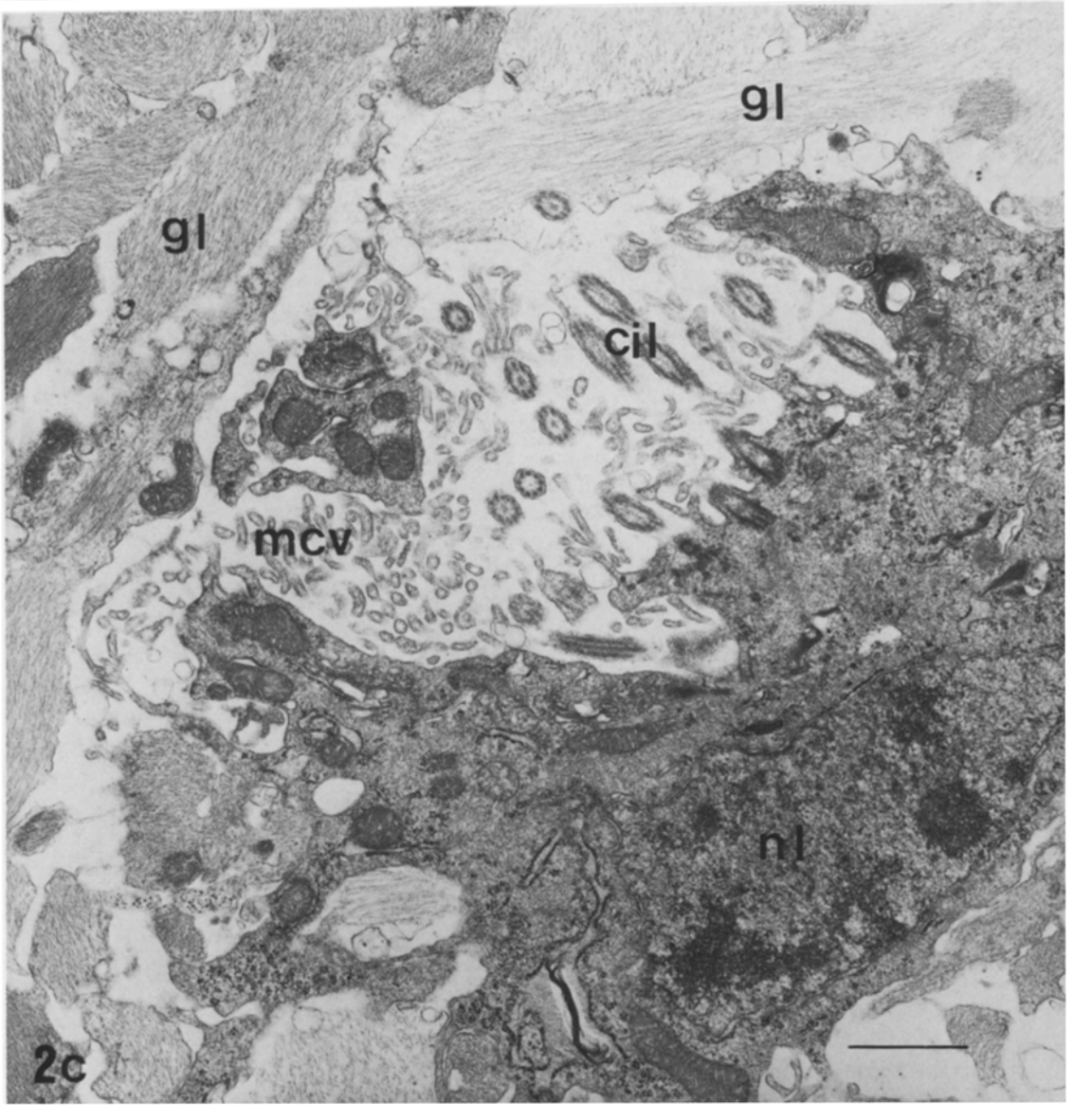
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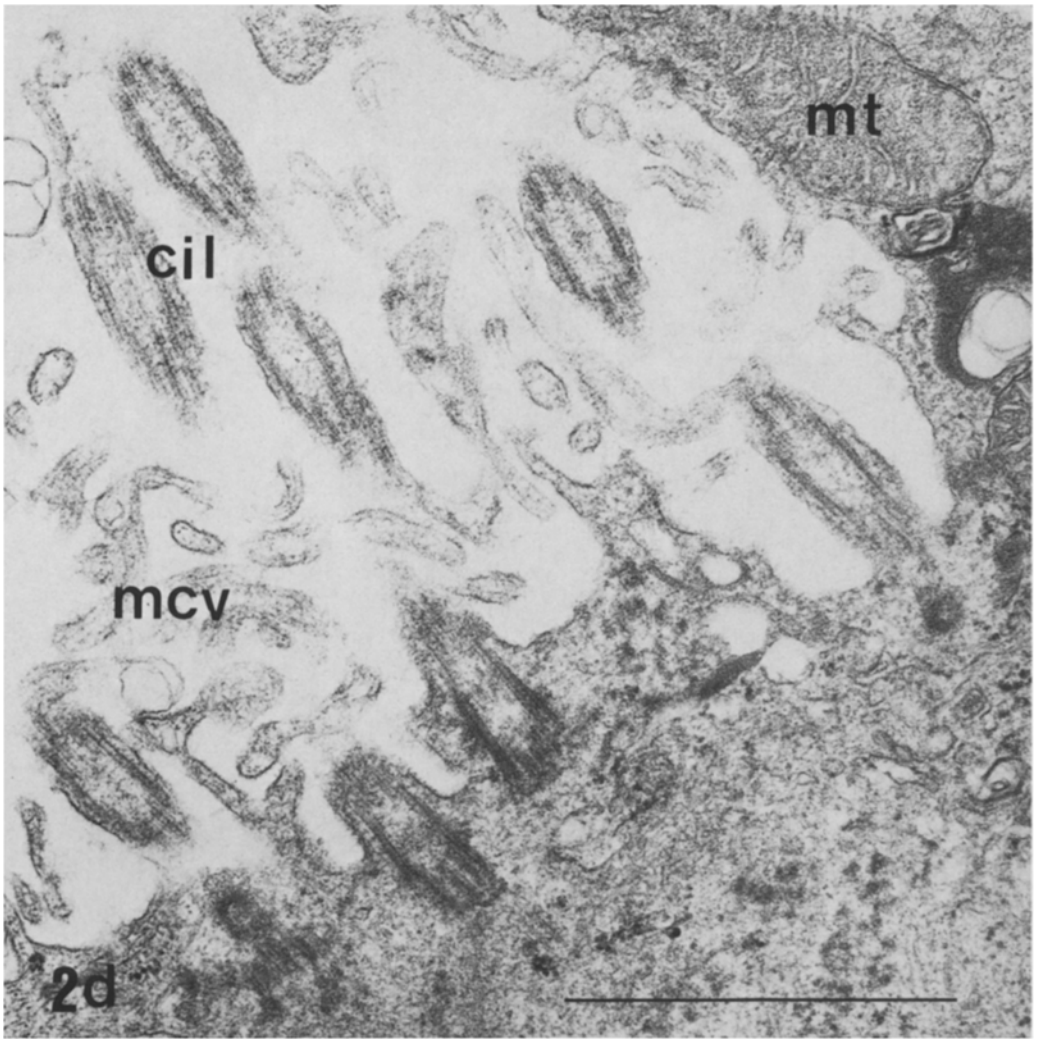

Fig. 2a–d. Electron microscopic observations of the wall of the Dandy-Walker cyst. **a** The outer surface of the cyst wall is covered by the tiers of arachnoid cells. *du* (dural side), *col* (collagenous tissue), *nl* (nucleus of the arachnoid cell). × 9,000. Bar: 1 μm. **b** The innermost surface of the cyst wall is covered by a layer of flattened cells (two *arrow-heads*) devoid of characteristic ependymal components. *gl* (neuroglial fibrils), *lu* (lumen of the cyst), × 9000. Bar: 1 μm. **c** A buried island of ependymal cells is seen in an intermediate layer of neuroglial tissue. *gl* (neuroglial fibrils), *cil* (cila), *mcv* (microvilli), and *nl* (nucleus) of the ependymal cells. × 6,300. Bar: 1 μm. **d** A high magnification of (**c**). The cilia (*cil*) in groups on the irregular apical surface of the ependymal cell and the microvilli (*mcv*) are also seen. *mt* (mitochondria of the ependymal cell). × 15,000. Bar: 1 μm

enlargement and deepening of the posterior fossa associated with elevation of the groove for the lateral sinus and torcula. A CT scan revealed hydrocephalus and a large cystic dilatation of the posterior fossa. The cerebellar hemispheres were symmetrically underdeveloped and were displaced laterally and anteriorly. Hypoplasia of the cerebellar vermis was also noted (Fig. 1). A vertebral angiogram demonstrated a posterior avascular mass, displaced posterior and inferior cerebellar arteries, and an elongated vein of Galen. These findings are highly compatible with a Dandy-Walker cyst. Though a right ventriculo-peritoneal shunt was performed on September 8, 1982, a marked posterior fossa cyst and symmetric enlargement of the third and lateral ventricles were still present postoperatively. On November 8, 1982, a suboccipital craniectomy was performed. When the dura was opened, the posterior fossa was found to be occupied by a large cyst, covered by a translucent membrane. A large portion of the membrane was removed but no attempt was made to remove it in its entirety. It could then be observed that the cerebellar hemispheres were widely separated (cerebellum bifidum) and that the hypoplastic vermis was situated anteriorly. The cyst was actually the greatly dilated fourth ventricle. The postoperative course was uneventful except for fever caused by pharyngitis. A postoperative CT scan showed slightly decreased third and lateral ventricles, but no change in the dilated fourth ventricle.

The surgical specimen of the excised cysts wall was examined by light and electron microscopy.

Materials and Methods

The excised cyst wall obtained during surgery was fixed immediately by immersion in could 2% glutaraldehyde in a 0.1 M sodium

cacodylate buffer (pH 7.4) overnight and then postfixed in 1 % OsO_4 for 60 min. It was dehydrated in graded concentrations of ethanol and then propylene oxide, and embedded in Epon 812. Ultrathin sections were stained with uranylacetate and lead citrate, and observed in a Hitachi HU-12A electron microscope.

For light microscopy, the cyst wall was fixed in formalin and embedded in paraffin and then stained by hematoxylin and eosin (HE).

Results

Light Microscopy

The cyst wall was composed of an outer layer of arachnoid tissue and an inner layer of mixed fibrous and neuroglial tissue.

Electron Microscopy

The outer surface of the cyst wall was covered by tiers of hyperplastic arachnoid cells, the outermost tiers of which were thicker and denser than the inner ones. The loosely arranged arachnoid cells were surrounded by collagen. The tortuous intercellular spaces preserved a certain gap (Fig. 2a). Beneath the arachnoid structures, the cerebellar structures were not completely lost, and clusters of neuroglial fibrils were loosely distributed despite the lack of neuronal tissues. However, the innermost surface of the cyst wall was generally covered by a layer of thin, flattened cells which were apparently devoid of ependymal cell components (Fig. 2b).

In the layer of the neuroglial fibril clusters, a small buried island of ependymal cells was seen. They lacked the typical cuboidal appearance, were bordered on one side by a lumen and by a strand of neuroglial fibrils on the basal surface. The irregular luminal surface was characterized by microvilli and cilia. The former were less numerous than those of the normal ventricular ependymal layer. The latter were scattered in groups on the luminal surface, and were straight and relatively stubby. These cilia displayed the usual pattern of 9 + 2 fibers structure. The intercellular spaces were tortuously arranged; there was apparently no true tight junction on the luminal surface. The basal surface abutted directly on the strand of neuroglial fibrils without any intervening basement membrane. The cytoplasm contained a somewhat irregular but ovoid nucleus, a few mitochondria, and relatively well developed rough-endoplasmic reticulum (Fig. 2c, d).

Discussion

Though various neuroradiologic examinations, including CT scan, have been very helpful in elucidating that the differentiation between a Dandy-Walker cyst and an arachnoid cyst or an enlarged cisterna magna is based on the fact that the former is intra-axial, confined within the fourth ventricle, and associated with agenesis or hypoplasia of the cerebellar vermis, whereas the latter are extra-axial and dorsal to the fourth ventricle, the most definitive diagnosis depends on the pathologic features of the cyst wall.

There have been several reports on light-microscopic observations of the walls of Dandy-Walker cysts [2, 4, 6–8, 10]. Most cyst walls consisted of an arachnoid cell layer on the outer surface, an ependymal cell layer on the inner surface, and varying amounts of neuroglial tissue.

Hart et al. [7] noted that Dandy-Walker cysts varied considerably in size and were composed of an outer layer of pia-arachnoid and an inner layer of ependyma in 21 cases, 12 of which demonstrated unequivocal cerebellar tissue between the ependyma and pia-arachnoid, usually near the cerebellar reflection, but occasionally further out along the membrane. They also found focal pressure atrophy of the ependyma in most cases, with focal gliosis in many and calcification in two instances. According to Carmel et al. [2], in five cases of autopsy findings of Dandy-Walker syndrome, the cyst walls of all cases demonstrated an outer pia-arachnoid layer and an inner ependymal layer; and clusters of cerebellar tissue, containing cells that were clearly cerebellar in origin, were found in the cyst walls in four cases.

The ultrastructure of the wall of Dandy-Walker cysts has rarely been reported, unlike that of arachnoid cysts [5, 9, 12, 14]. The cyst wall in our case, consisted of an outer arachnoid cell layer similar to the wall of an arachnoid cyst, interwoven neuroglial strands of cerebellar origin, a small, buried island of ependyma, and an innermost cell layer lacking the characteristic appearance of the ependyma. It is very interesting that this small isolated and buried island of ependymal cells was seen in a layer of neuroglial tissue. Gibson [4] also described light-microscopically a similar small buried island of ependyma in a layer of white matter in the wall of a Dandy-Walker cyst.

This unusual ultrastructural finding in the wall of the Dandy-Walker cyst may result from degradation in the wall or long-standing pressure atrophy of the ependyma. To understand the possible pathogenesis of the Dandy-Walker syndrome, it may be necessary to concentrate on the ultrastructure of the wall of the Dandy-Walker cyst.

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